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Critical Review of Infantile Fibrous Dysplasia: Surgical Treatment

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Summary: A critical review of the surgical treatment of 65 patients with infantile fibrous dysplasia demonstrated that "circumscribed" types of the disease generally do not require surgical treatment, while "extended" types, as well as Albright's syndrome, require early surgical treatment aimed at preventing development of skeletal deformities which are difficult to correct later. Prophylactic intramedullary nailing with nails of suitable caliber is most effective. **Key Words:** Curettage—Fibrous dysplasia—Intramedullary nailing—Rigid osteosynthesis.

During growth, localization of fibrous dysplasia in the tubular bones, particularly in "extended" types of the disease and in Albright's syndrome, may lead to progressive and often severe skeletal deformity as well as to pathologic fractures (3,6,9). In addition to sarcomatous degeneration, which is rare (2), these mechanical complications constitute the principal indications for treatment of this dysplasia which, at the end of growth, loses its potential to proliferate and "matures" into fibrous osseous tissue (1,4). Some investigators (6,7,12) have proposed surgical treatment to prevent mechanical complications. We critically reviewed the operative treatment of 65 patients with fibrous dysplasia, particularly in relation to the results obtained by different treatments.

PATIENTS AND METHODS

The clinical and radiographic data of 65 patients (38 males and 27 females) with an average age of 8.6 years (range 1–16 years) at onset of treatment were analyzed. Histologic diagnosis was made and operations were performed at the Istituto Ortopedico Rizzoli between 1956 and 1986.

Two different skeletal sites in three patients were treated. Sixty-eight local sites were operatively treated (1 pelvis, 45 femur, 16 tibia, 5 humerus, 1 forearm). Thirty-one patients were operated on more than once (average three times), and 124 operations were performed (Table 1), with an average follow-up of 6.5 years (range 6 months to 27 years).

TABLE 1. *Operative treatment*

Procedure	n
Emptying and curettage	83
Rigid osteosynthesis	31
Intramedullary nailing	8
Resection	2
Total	124

The cases were divided into "circumscribed" and "extended" types of the disease (Fig. 1). We defined a "circumscribed" fibrous dysplasia as any lesion, on radiograph, less than one fourth of the bone segment and only one cortex involved (32 cases monostotic and five polyostotic); an "extended" fibrous dysplasia is any lesion involving more of the bone segment and both hemidiaphyses (22 polyostotic and six monostotic cases).

In this series, the indications for operation differed considerably for each type and the results were considered separately. Surgical treatment of the circumscribed forms (37 patients) was limited in 35 patients to incisional biopsy and curettage. In

TABLE 2. *Circumscribed forms of infantile fibrous dysplasia (37 patients)*

		Results	
Operative treatment	n	Positive	Negative
Emptying and curettage	38	16 (42%)	22 (58%)
Resection	2	1 (50%)	1 (50%)

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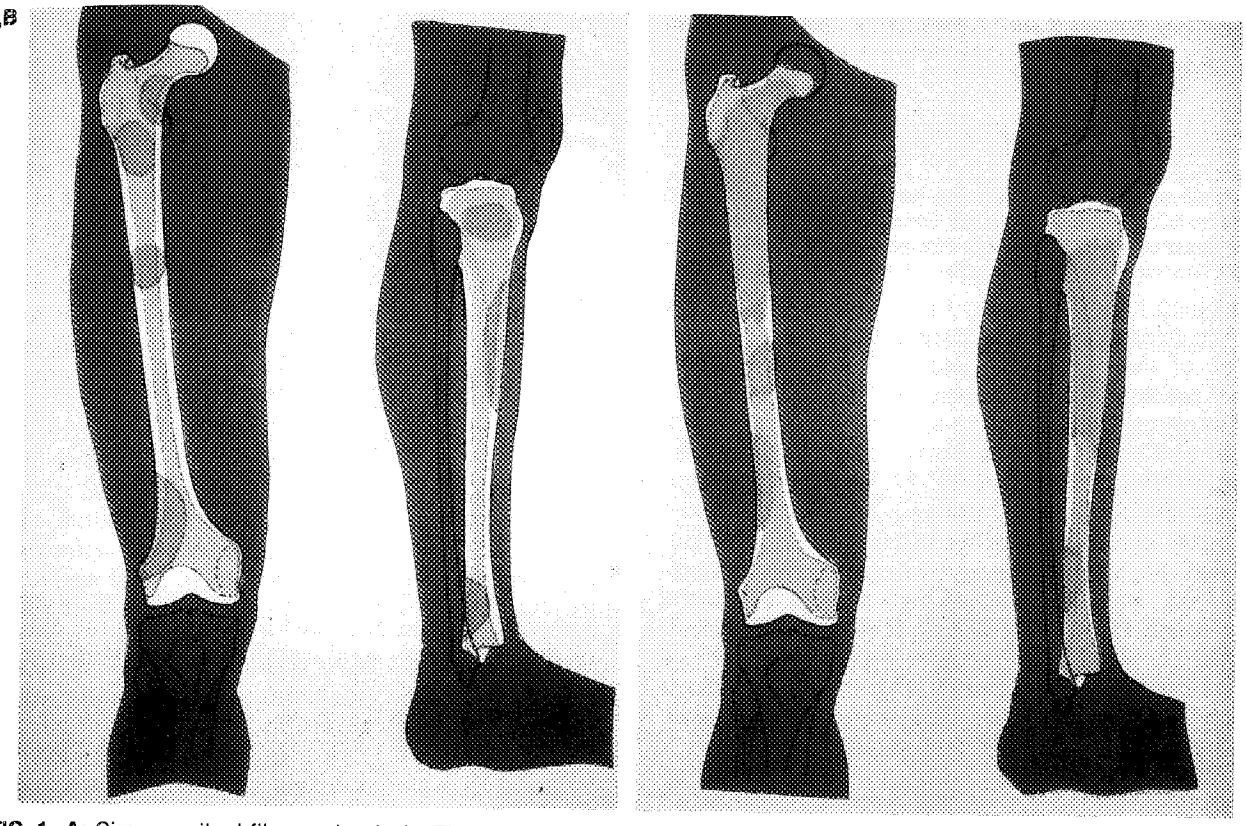

FIG. 1. A: Circumscribed fibrous dysplasia. The lesion involves less than one-fourth of the entire bone segment and only one cortex. **B:** Extended fibrous dysplasia. The lesion involves one-fourth of the entire bone segment and both hemidiaphyses.

two patients with a femoral diaphysis had diaphyseal hemiresection. In the "extended" forms and in Albright's syndrome (84 sites operated on in 28 patients) the indications for operation were based on mechanical complications (deformity and pathologic fracture, often simultaneous) and were aimed at correction or prevention of progression. Many of these lesions required subsequent operations. Methods used were emptying and curettage, osteosynthesis with plate and screws, and intramedullary osteosynthesis. The result of the surgical procedure was judged positive if the dysplastic area healed or mechanical complications were prevented.

RESULTS

Emptying and curettage led to negative results in ~60% of patients with circumscribed dysplasia.

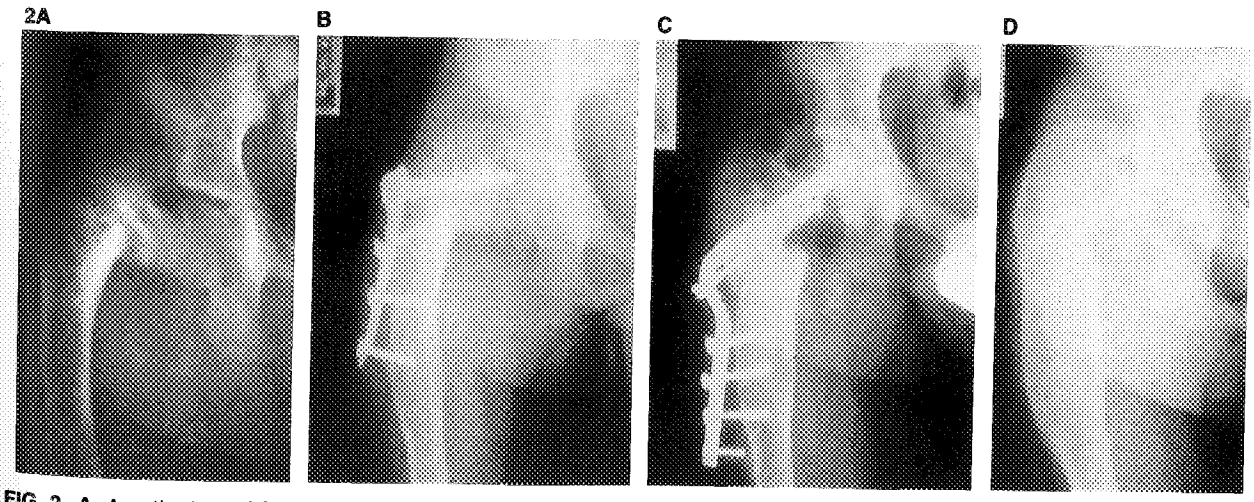

FIG. 2. A: A patient aged 9 years with extended fibrous dysplasia and severe coxa vara previously underwent three successive emptyings and curettages. **B and C:** Two osteotomies were performed, resulting in loosening of the fixation. **D:** Two years after removal, the deformity progressed. Results were negative.

TABLE 3. *Extended forms of infantile fibrous dysplasia and Albright's syndrome (28 patients)*

Operative treatment	n	Results	
		Positive	Negative
Emptying and curettage	45	0	45 (100%)
Rigid osteosynthesis	31	0	31 (100%)
Intramedullary nailing	8	6 (75%)	2 (25%)

This high failure rate may be attributed exclusively to the persistence of an osteolytic area; however, none of these children later had pathologic fractures or severe bony deformities (Table 2).

Emptying and curettage and osteosynthesis with plate and screws did not produce positive results (Fig. 2) in extended forms and in Albright's syndrome (Table 3), but intramedullary fixation gave negative results in only two of eight children (25%). One of the two had a progressive deformity postoperatively, probably caused by inadequate fixation (Rush nail). The other had undergone femoral nailing by the Kuntscher method, performed after five other operations had failed. The patient had postoperative infection and, after removal of the nail, severe limb length discrepancy remained.

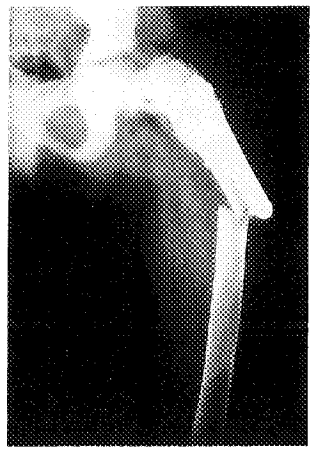

FIG. 3. Fracture at the plate apex.

DISCUSSION

Our disappointing results show that in infantile fibrous dysplasia one must first establish the extent of the dysplastic area and determine whether it is circumscribed or extended.

In circumscribed fibrous dysplasia, because the lesion is minimally aggressive, monitoring progres-

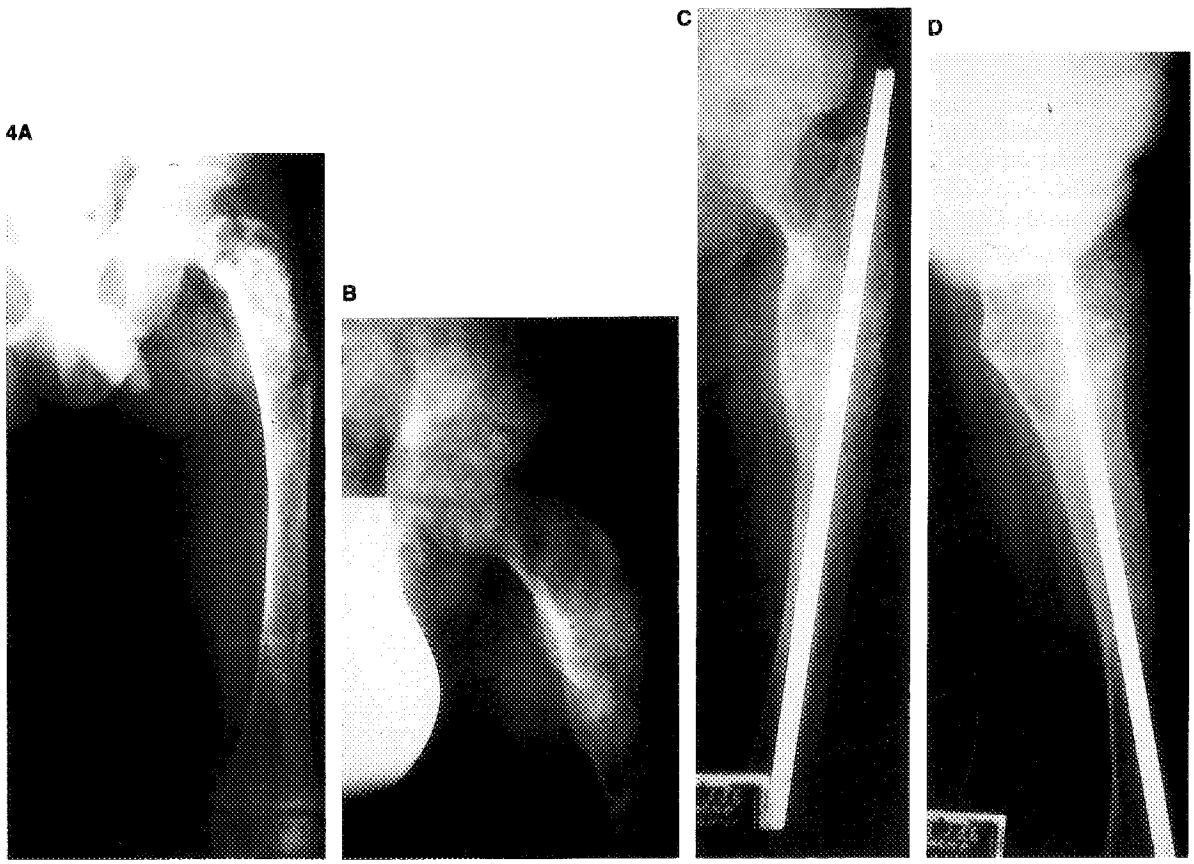

FIG. 4. **A:** Example of extended fibrous dysplasia showing fractures and varus deformity. **B:** Valgus osteotomy with rigid osteosynthesis was performed because of plate loosening and recurrence of the deformity. **C and D:** After 7 years, further corrective osteotomy and intramedullary nailing were performed. Follow-up result after 1 year was positive.

sion is sufficient; surgical intervention is necessary only when complications occur. When diagnosis is dubious, biopsy is necessary and must be performed according to the traditional procedure with emptying and curettage only after diagnosis has been histologically confirmed.

On the contrary, in extended forms of the disease and in Albright's syndrome, early intervention may be necessary before deformity occurs, because such conditions are extremely difficult to correct surgically later. In such cases, emptying and curettage should be avoided because it neither heals the osteolytic area nor modifies the aggressive progression of the disease.

Correction and prevention of the deformity by rigid fixation with compression plates failed consistently. Such treatment was frequently associated with mobilization of the plates, recurrence of the deformity, or bone fracture at the lower margin of the plate (Fig. 3) owing to the poor properties of pathologic bone or to concentration of tension of the bone plate interface.

Of the various methods used in this series, intramedullary nailing undoubtedly produced the best results (Fig. 4), thus confirming the experience reported by other investigators (5,6,8,10–12). We believe that the relatively high percentage of negative results reported may be prevented by using nails of a suitable caliber and by intervening early, i.e., before the patient has had to undergo repeated operations and before severe deformity occurs.

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